# ANGIOLYMPHOID HYPERPLASIA WITH EOSINOPHILIA- A CASE REPORT

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ABSTRACT : *Angiolymphoid hyperplasia with eosinophilia ( ALHE ) is a benign condition characterized by subcutaneous lesions in the head-neck region. It is frequently misdiagnosed as a malignant lesion. Knowledge of the existence of the disease and pathological interpretation are requisites for early diagnosis. We present a case report and review of the literature.*

## INTRODUCTION

In 1969 Wells and Whimster described nine patients in a dermatology clinic with subcutaneous nodules in the head – neck region with occasional regional lymph node enlargements They called it subcutaneous angioblastic lymphoid hyperplasia with eosinophilia Until the late 1970s, it was believed to be synonymous with and equivalent to Kimura's disease, which also causes angiomatous lesions, lymphadenopathy and marked eosinophilia Subsequent studies ( Rosai et al,1979, Googe et al, 1987) indicated that these are two separate entities with different clinical and histological features Due to the predilection of ALHE for the head-neck region, all surgeons treating conditions in this region should be aware of the clinical features and management options available

## CASE REPORT

A 20 year old male was brought to our outpatient department with multiple bilateral neck swellings of 3 to 6 years duration There was history of surgery and excision biopsy of one of the swellings done elsewhere The biopsy report indicated non-specific inflammation On examination, multiple swellings ranging in size from 2cm to 4cm diameter, were noted in the post auricular and upper cervical region bilaterally The swellings were non tender, firm and had a smooth surface There were no palpable lymph nodes Blood cell count did not reveal eosinophilia Fine needle aspiration cytology of the swellings was inconclusive Our clinical impression was of a soft tissue malignancy An excision biopsy was performed under general anesthesia The masses were found to be discrete from each other as well as from the underlying tissues They were fibrotic, well-vascularised and lying in the subcutaneous tissue Histological examination showed ( Fig -I ) vascularised connective tissue with proliferation of small blood vessels The blood vessels were lined by plump endothelial cells and surrounded by a number of eosinophils and a lymphoid infiltrate with formation of germinal centres There was no evidence of hyperchromatism, mitotic activity or malignancy in all the sections studied Thus a diagnosis of ALHE was established

## DISCUSSION

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a rare condition causing nodular or papular angiomatous lesions in the dermis , subcutaneous tissues and adjacent lymphnodes Although benign, the disease tends to persist, grows slowly and is of unknown etiology It is commonly found in the head- neck region although reports suggest (Smith et al,1988, Akosa et al, 1990) that it can also occur in the orbit, arm, oral mucosa, or even in the parotid gland Amongst Oriental patients it is predominantly seen in young (second decade) males(85%) while it tends to occur in females (70%) and older patients (third or fourth decades) amongst Caucasians (Henryet al,1978) Histologically, ALHE is characterized by proliferation of small to medium sized blood vessels often with lobular architecture Many of these blood vessels are lined by plump, enlarged endothelial cells causing occlusive changes in the arteries This results in the typical "cobblestone" appearance The perivascular infiltrate consists of lymphocytes and eosinophils Generally, eosinophils form 5 to 15 % of the cells and a complete absence of eosinophils makes the diagnosis of ALHE doubtful Until the late 1970's, ALHE was thought to be equivalent to and synonymous with Kimura's disease, a disease prevalent in

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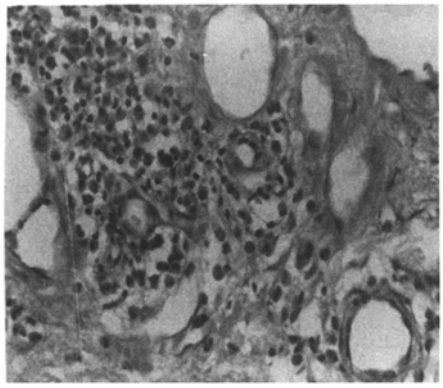

Fig I ALHE H & E Stain 20X showing eosinophils & proliferative blood vessels lined by plump endothelial cells

Japan, China and South Asia The appearance of lesions in ALHE and Kimura's disease is similar with a raised erythematous skin lesion or subcutaneous nodule being common Histologically, both have similar features like extensive lymphoid proliferation, tissue eosinophilia and vascular hyperplasia The differences in Kimura's disease are that the vascular hyperplasia is less exuberant, the newly formed blood vessels are canalized and lined by flat endothelial cells (Rosai et al,1979)(See Table 1) Other points of difference seen in Kimura's disease are a wider age span, male predominance, a tendency for extensive lesions, often with involvement of salivary tissue and lymph nodes and at sites distant from the head -neck region Some authors (Rosai et al ,1979, Googe et al ,1987) believe that ALHE represents a stage of histiocytoid or epithelial haemangio-ma, which is a true vascular neoplasm while Kimura's disease is a localized manifestation of a systemic immunological reaction Other authors (Akosa et al, 1990) disagree and believe that ALHE represents an atopic reaction to a variety of agents

**Table I**

Diferences between ALHE and Kumura's disease

| **Features** | **ALHE** | **Kimura's** |
|---|---|---|
| Location | Mainly head – neck area | Generalized |
| Lympha-denopathy | Local Rare | Systemic Marked |
| Blood eosinophil | Numerous new | |
| Histology | blood vessels lined by plump endo-thelial cells | New vessels lined by Flat endothe lial cells |

The appearances of the lesion in ALHE can be mistaken for angiosarcoma, haemangioendothelioma ,malignant lymphoma, Kaposi's sarcoma, haemangioma or dermatofibroma We experienced this difficulty in diagnosis The differences are histological in angiosarcoma eosinophils are rarely present, while nuclear atypia , hyperchromatism , mitotic activity and dissection of collagen are its distinctive features, in reticular haemangioendothelioma a reticular growth pattern with tall, narrow endothelial cells and a typical hob-nail appearance are seen , in most benign angiomas or ectasias, an inflammatory cell infiltrate, as seen in ALHE, is absent Therefore, knowledge of the existence of the disease and its histological features is very important Accurate clinical and cytological diagnosis of ALHE allows for initial treatment with intralesional or systemic steroids (Ingrams et al, 1995) although local excision with a safe margin of healthy tissue is the treatment of choice Insufficient removal can result in recurrence In residual or recurrent masses use of laser has been advocated to reduce hemorrhage (Vallis et al, 1988)

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